General

Guideline Title

An official American Thoracic Society clinical practice guideline: diagnosis, risk stratification, and management of pulmonary hypertension of sickle cell disease.

Bibliographic Source(s)

Klings ES, Machado RF, Barst RJ, Morris CR, Mubarak KK, Gordeuk VR, Kato GJ, Ataga KI, Gibbs JS, Castro O, Rosenzweig EB, Sood N, Hsu L, Wilson KC, Telen MJ, DeCastro LM, Krishnamurti L, Steinberg MH, Badesch DB, Gladwin MT, American Thoracic Society Ad Hoc Committee on Pulmonary Hypertension of Sickle Cell Disease. An official American Thoracic Society clinical practice guideline: diagnosis, risk stratification, and management of pulmonary hypertension of sickle cell disease. Am J Respir Crit Care Med. 2014 Mar 15;189(6):727-40. [75 references] PubMed

Guideline Status

This is the current release of the guideline.

This guideline meets NGC's 2013 (revised) inclusion criteria.

Recommendations

Major Recommendations

Definitions for the levels of evidence (high, moderate, low, very low) and the strength of the recommendations (strong, weak) are provided at the end of the "Major Recommendations" field.

Estimating Mortality Risk in Sickle Cell Disease (SCD)

Conclusions from the Committee:

- Risk stratification guides decision making.
- Mortality risk can be determined by noninvasive measurement of the tricuspid regurgitant velocity (TRV) via Doppler echocardiography.
- Serum N-terminal pro-brain natriuretic peptide (NT-pro-BNP) measurement is a reasonable noninvasive alternative if Doppler echocardiography is either unavailable or cannot obtain adequate images. (*Note*: Measurements may be misleading in patients with renal insufficiency.)
- Mortality risk can also be determined invasively by direct hemodynamic measurements via right heart catheterization (RHC).

Treatment of Patients with SCD Who Have Increased Mortality Risk

Hydroxyurea

Recommendation: For patients with SCD who have an increased risk for mortality, the Committee recommends hydroxyurea therapy (*strong recommendation, moderate-quality evidence*).

Remarks: The Committee defines an increased risk for mortality as a TRV of ≥2.5 m/second, an NT-pro-BNP level of ≥160 pg/ml, or RHC-confirmed pulmonary hypertension (PH).

Values and preferences: This recommendation attaches a relatively high value to the potential benefits from therapy and a lower value to the risks and burdens of therapy.

Chronic Transfusion Therapy

Recommendation: For patients with SCD who have an increased risk for mortality and who are either not responsive to hydroxyurea or are not candidates for hydroxyurea, the Committee suggests chronic transfusion therapy (*weak recommendation, low-quality evidence*).

Remarks: The Committee defines an increased risk for mortality as a TRV of ≥2.5 m/second, an NT-pro-BNP level of ≥160 pg/ml, or RHC-confirmed PH.

Chronic Anticoagulant Therapy

Recommendation: For patients with SCD who have RHC-confirmed PH, venous thromboembolism, and no additional risk factors for bleeding, the Committee suggests indefinite anticoagulant therapy rather than a limited duration of therapy (*weak recommendation, low-quality evidence*).

Values and preferences: This recommendation attaches a relatively high value to the prevention of recurrent venous thromboembolic events and a lower value to the risks and burdens of anticoagulant therapy.

Targeted Pulmonary Arterial Hypertension (PAH) Therapy

Recommendation: For all patients with SCD who have elevated TRV alone, or elevated NT-pro-BNP alone, and for most patients with SCD who have RHC-confirmed PH, the Committee recommends *against* targeted PAH therapy (*strong recommendation, moderate-quality evidence*).

Remarks: Targeted PAH therapy refers to prostacyclin agonist, endothelin receptor antagonist, soluble guanylate cyclase stimulator, and phosphodiesterase-5 inhibitor therapy.

Values and preferences: This recommendation places a greater value on avoiding expensive and potentially harmful agents in patients who may not benefit and a lower value on avoiding the complications of invasive hemodynamic testing by using noninvasive results for decision making.

Recommendation: For select patients with SCD who have an RHC-confirmed marked elevation of their pulmonary vascular resistance, normal pulmonary artery wedge pressure, and related symptoms, the Committee suggests a trial with either a prostacyclin agonist or an endothelin receptor antagonist (*weak recommendation, low-quality evidence*).

Remarks: This group of patients with SCD is characterized by a mean pulmonary arterial pressure (mPAP) of ≥25 mm Hg with a pulmonary artery wedge pressure <15 mm Hg, plus increased pulmonary vascular resistance (PVR) of ≥160 dyn · seconds · cm^{-5} (2 Wood units). Patients with SCD who have confirmed PH are now categorized as being within World Health Organization (WHO) group 5, instead of WHO group 1. As a result, use of targeted PAH therapy in such patients is considered off-label.

Values and preferences: This recommendation places a greater value on improving symptoms and hemodynamic measures and a lower value on the costs, burdens, and risks of therapy.

Recommendation: For patients with SCD who have an RHC-confirmed marked elevation of their pulmonary vascular resistance, a normal pulmonary artery wedge pressure, and related symptoms, the Committee recommends *against* phosphodiesterase-5 inhibitor therapy as a first-line treatment (*strong recommendation, moderate-quality evidence*).

Remarks: The recommendation is based on the observation that phosphodiesterase-5 inhibitor therapy may increase the risk of hospitalization for vasoocclusive crisis.

Values and preferences: This recommendation places a higher value on avoiding serious complications and a lower value on symptomatic and hemodynamic improvement.

Definitions:

Levels of Evidence

The identified evidence was appraised using the Grading of Recommendations Assessment, Development and Evaluation (GRADE) approach. In the four level grading system (high, moderate, low, and very low quality evidence), randomized trials begin with the assumption of high quality evidence, whereas well-conducted observational studies (e.g., cohort studies, case-control studies) begin with an assumption of low quality evidence. The quality of evidence can be downgraded or upgraded on the basis of predefined criteria.

Strength of Recommendations

The decision of whether to recommend an intervention was made by consensus and based upon four criteria: the quality of evidence, the balance of desirable and undesirable consequences, patient values and preferences related to the intervention and outcomes, and resource use.

A strong recommendation was made when there was certainty about the balance of desirable and undesirable consequences of an intervention, whereas a weak recommendation was made when there was less certainty or the balance of desirable and undesirable consequences was finely balanced.

Clinical Algorithm(s)

An algorithm titled "Proposed algorithm for evaluation of pulmonary hypertension related to sickle cell disease" is provided in the original guideline document.

Scope

Disease/Condition(s)

Pulmonary hypertension of sickle cell disease (SCD)

Guideline Category

Diagnosis

Evaluation

Management

Risk Assessment

Treatment

Clinical Specialty

Cardiology

Critical Care

Family Practice

Hematology

Internal Medicine

Pediatrics

Pulmonary Medicine

Intended Users

Advanced Practice Nurses

Nurses

Physicians

Guideline Objective(s)

- To provide the basis for rational decisions in the management of patients with sickle cell disease (SCD) who have an increased risk for death
- To advise hematologists, pulmonologists, cardiologists, pediatricians, and internists about how to identify and manage patients with SCD who are at increased risk for mortality

Target Population

Adult and pediatric patients with sickle cell disease (SCD) who are at increased risk for mortality

Interventions and Practices Considered

Risk Assessment/Diagnosis

1. Noninvasive measurement of the tricuspid regurgitant velocity (TRV) via Doppler echocardiography
2. Serum N-terminal pro-brain natriuretic peptide (NT-pro-BNP) measurement
3. Direct hemodynamic measurements via right heart catheterization (RHC)

Management/Treatment

1. Hydroxyurea
2. Chronic transfusion therapy
3. Chronic anticoagulant therapy
4. Targeted pulmonary arterial hypertension (PAH) therapy: prostacyclin agonist, endothelin receptor antagonist, soluble guanylate cyclase stimulator, and phosphodiesterase-5 inhibitor therapy

Major Outcomes Considered

- Mortality risk
- Adverse effects
- Other patient-important outcomes
    - Quality of life
    - Hospitalization for vasoocclusive crisis or acute chest syndrome
    - Stroke
    - Acute venous thromboembolism
    - Exercise capacity
    - Functional status
    - Cardiopulmonary hemodynamics

Methodology

Methods Used to Collect/Select the Evidence

Searches of Electronic Databases

Description of Methods Used to Collect/Select the Evidence

To identify evidence relevant to each clinical question, systematic reviews of the literature were performed. The pre-specified search strategies, including search dates and terms, are described in Tables E6 to E12 in the online supplement appendix (see the "Availability of Companion Documents" field). The systematic reviews were periodically updated and are current through February 15, 2013. Databases searched included MEDLINE (PubMed), Cochrane Controlled Clinical Trials (CENTRAL), and the Cochrane Database of Systematic Reviews.

Study Selection Criteria for the Use of the Tricuspid Regurgitant Velocity (TRV) to Assess Mortality Risk in Patients with Sickle Cell Disease (SCD)

Studies were selected if they (a) enrolled patients with SCD, (b) compared patients who had their TRV measured via Doppler echocardiography to patients who did not have their TRV measured, and (c) measured patient-important outcomes. Alternatively, studies were selected if they measured the mortality risk among patients who had an elevated TRV, relative to those who had a normal TRV.

Study Selection Criteria for the Use of the N-terminal Brain Natriuretic Peptide (NT-pro-BNP) to Assess Mortality Risk in Adults with SCD

Studies were selected if they (a) enrolled patients with SCD, (b) compared patients who had their NT-pro-BNP level measured to patients who did not have their NT-pro-BNP level measured, and (c) measured patient-important outcomes. Alternatively, studies were selected if they measured the mortality risk among patients who had an elevated NT-pro-BNP level, relative to those who had a normal NT-pro-BNP level.

Study Selection Criteria for the Use of Hydroxyurea in Patients with SCD and Either Right Heart Catheterization (RHC)-Confirmed Pulmonary Hypertension, a TRV ≥2.5 m/s, or an NT-Pro-BNP Level ≥160 pg/ml

Studies were selected if they (a) enrolled patients with SCD, (b) compared patients who received hydroxyurea to patients who received no therapy or placebo, and (c) measured patient-important outcomes. Given the anticipated large volume of evidence, the guideline committee initially sought published systematic reviews that included trials that met these selection criteria, with the plan to search step-wise for randomized trials and then observational studies if no suitable systematic reviews were identified. If such systematic reviews were identified, it was planned to combine the systematic review with relevant studies published after the systematic review. Studies identified in this fashion were to be supplemented with unsystematic observations from the committee members.

Study Selection Criteria for the Use of Chronic Transfusion Therapy in Patients with SCD and Either RHC-Confirmed Pulmonary Hypertension, a TRV ≥2.5 m/s, or an NT-pro-BNP Level ≥160 pg/ml

Studies were selected if they (a) enrolled patients with SCD, (b) compared patients who received chronic transfusion therapy to patients who received no chronic transfusion therapy, and (c) measured patient-important outcomes. Given the anticipated large volume of evidence, the guideline committee initially sought published systematic reviews that included trials that met these selection criteria, with the plan to search step-wise for randomized trials and then observational studies if no suitable systematic reviews were identified. If such systematic reviews were identified, it was planned to combine the systematic review with relevant studies published after the systematic review. Studies identified in this fashion were to be supplemented with unsystematic observations from the committee members.

Study Selection Criteria for the Use of Lifelong Anticoagulant Therapy in Patients with SCD-Related Pulmonary Hypertension and Thromboembolic Disease

Studies were selected if they (a) enrolled patients with SCD-related pulmonary hypertension and venous thromboembolic disease, (b) compared patients who received lifelong anticoagulant therapy to patients who received a shorter duration of anticoagulant therapy, and (c) measured patient-important outcomes.

Study Selection Criteria for the Use of Lifelong Anticoagulant Therapy in Patients with SCD and Thromboembolic Disease

â€‹Studies were selected if they (a) enrolled patients with SCD and venous thromboembolic disease, (b) compared patients who received lifelong anticoagulant therapy to patients who received a shorter duration of anticoagulant therapy, and (c) measured patient-important outcomes.

Study Selection Criteria for the Use of Targeted Pulmonary Arterial Hypertension (PAH) Therapy in Patients with SCD

Studies were selected if they (a) enrolled patients with SCD-related pulmonary hypertension, (b) compared patients who received targeted PAH therapy to patients who received no targeted PAH therapy, and (c) measured patient-important outcomes.

Number of Source Documents

Refer to Figures E1 though E5 in the online supplement (see the "Availability of Companion Documents" field) for study selection flow charts.

Methods Used to Assess the Quality and Strength of the Evidence

Weighting According to a Rating Scheme (Scheme Given)

Rating Scheme for the Strength of the Evidence

Levels of Evidence

The identified evidence was appraised using the Grading of Recommendations Assessment, Development and Evaluation (GRADE) approach. In the four level grading system (high, moderate, low, and very low quality evidence), randomized trials begin with the assumption of high quality evidence, whereas well-conducted observational studies (e.g., cohort studies, case-control studies) begin with an assumption of low quality evidence. The quality of evidence can be downgraded or upgraded on the basis of predefined criteria.

Methods Used to Analyze the Evidence

Review of Published Meta-Analyses

Systematic Review with Evidence Tables

Description of the Methods Used to Analyze the Evidence

Evidence tables were constructed using GRADEpro. Refer to Tables E1 through E5 in the online supplement of the guideline (see the "Availability of Companion Documents" field).

Methods Used to Formulate the Recommendations

Expert Consensus

Description of Methods Used to Formulate the Recommendations

Committee Composition

The project chair and co-chairs selected 24 committee members based upon their expertise in sickle cell disease, pulmonary hypertension, or both. The committee consisted of seven adult pulmonologists, six adult hematologists, four pediatric hematologists, two adult cardiologists, two pediatric cardiologists, one pediatric pulmonologist, one adult pulmonologist/methodologist and one pediatric emergency medicine physician.

Formulating Recommendations

Recommendations were formulated to answer the clinical questions created by the group. The decision of whether to recommend an intervention was made by consensus and based upon four criteria: the quality of evidence, the balance of desirable and undesirable consequences, patient values and preferences related to the intervention and outcomes, and resource use.

Sub-Committees

Two subcommittees were formed to identify important clinical questions related to: (1) screening and diagnosis and (2) treatment. These questions were subsequently discussed during an initial face-to-face meeting at the 2009 American Thoracic Society (ATS) International Conference. A writing committee was subsequently formed and divided into subgroups. Each subgroup was charged with preparing a portion of the guidelines, which entailed identifying, appraising, and synthesizing the evidence, as well as formulating recommendations and writing.

Document Preparation

The drafts from the various subgroups were integrated by the committee chairs. The document was distributed for discussion at two face-to-face meetings (the 2009 American Society of Hematology International Conference and the 2010 ATS International Conference) and periodically by e-mail and conference calls. All of the committee members had the opportunity to discuss and express any concerns about the document or the recommendations.

Rating Scheme for the Strength of the Recommendations

Strength of Recommendations

The decision of whether to recommend an intervention was made by consensus and based upon four criteria: the quality of evidence, the balance of desirable and undesirable consequences, patient values and preferences related to the intervention and outcomes, and resource use.

A strong recommendation was made when there was certainty about the balance of desirable and undesirable consequences of an intervention, whereas a weak recommendation was made when there was less certainty or the balance of desirable and undesirable consequences was finely balanced.

Cost Analysis

A formal cost analysis was not performed and published cost analyses were not reviewed.

Method of Guideline Validation

Internal Peer Review

Description of Method of Guideline Validation

The official clinical practice guideline of the American Thoracic Society (ATS) was approved by the ATS Board of Directors, November 2013.

Evidence Supporting the Recommendations

Type of Evidence Supporting the Recommendations

The type of supporting evidence is identified and graded for each recommendation (see "the Major Recommendations" field).

Benefits/Harms of Implementing the Guideline Recommendations

Potential Benefits

- Reduced variability and improved quality of care for patients with sickle cell disease (SCD) who are at increased risk for death
- Identifying patients at increased risk for mortality is important because it guides decision making. Specifically, it identifies patients who may benefit from SCD-specific therapies that have been shown to improve outcomes in patients with various complications of SCD, as illustrated previously with hydroxyurea. Risk stratification also allows patients to anticipate their future health care needs.

Refer to the original guideline document and the online supplement (see the "Availability of Companion Documents" field) for discussions of the balance of benefits versus harms of specific interventions recommended for the diagnosis and management of pulmonary hypertension.

Potential Harms

- In one study, mild to moderate neutropenia (i.e., an absolute neutrophil count of 500–1,249/mm^3) was significantly more common among

the HU group (46.9 vs. 18.6%; RR, 2.53; 95% CI, 1.58–4.03) than in the placebo group and required long-term dose reductions in nine patients. Severe neutropenia (i.e., absolute neutrophil count, 500/mm^3) was rare and not complicated by infection.

- Risks of chronic transfusion therapy include febrile reactions, allergic reactions, hemolytic reactions, and volume overload. Such adverse effects collectively occur in 15% of patients. Red cell alloimmunization is another consequence of chronic transfusion therapy, occurring in 11% of patients.
- The most common adverse effects during endothelin receptor antagonist therapy were headache (15%) and peripheral edema (21%); transaminase elevation was reported in 14% of patients.
- Indefinite anticoagulant therapy is associated with a 2.4% increased bleeding risk, increased cost, and the burden of monitoring.
- One placebo-controlled trial of sildenafil was prematurely discontinued because of an increase in serious adverse events in the sildenafil group, primarily hospitalization for pain.
- Renal dysfunction is independently associated with high N-terminal pro-brain natriuretic peptide (NT-pro-BNP) levels (15, 30) and, thus, testing is more likely to provide false-positive results in patients with renal dysfunction than in patients with normal renal function. In such patients, it is possible that the undesirable consequences of false-positive results (i.e., unnecessary therapy and the risks associated with it) might exceed the benefits of NT-pro-BNP testing. Until the impact of renal dysfunction on NT-pro-BNP levels is further studied, the committee suggests that NT-pro-BNP testing not be used in patients with renal dysfunction.

Refer to the original guideline document and the online supplement (see the "Availability of Companion Documents" field) for additional discussions of the balance of benefits versus harms of specific interventions recommended for the diagnosis and management of pulmonary hypertension.

Qualifying Statements

Qualifying Statements

These American Thoracic Society (ATS) guidelines about the diagnosis, risk stratification, and management of pulmonary hypertension of sickle cell disease (SCD-PH) are not intended to impose a standard of care. They provide the basis for rational decisions in the management of patients with SCD who have an increased risk for death, as defined by an elevated tricuspid regurgitant velocity (TRV), elevated serum N-terminal pro-brain natriuretic peptide (NT-pro-BNP) level, or right heart catheterization (RHC)-confirmed PH. Clinicians, patients, third-party payers, institutional review committees, other stakeholders, or the courts should never view these recommendations as dictates. No guidelines and recommendations can take into account all of the often compelling unique individual clinical circumstances. Therefore, no one charged with evaluating clinicians' actions should attempt to apply the recommendations from these guidelines by rote or in a blanket fashion. Statements about the underlying values and preferences as well as qualifying remarks accompanying each recommendation are its integral parts and serve to facilitate more accurate interpretation. They should never be omitted when quoting or translating recommendations from these guidelines.

Implementation of the Guideline

Description of Implementation Strategy

An implementation strategy was not provided.

Implementation Tools

Clinical Algorithm

For information about availability, see the *Availability of Companion Documents* and *Patient Resources* fields below.

Institute of Medicine (IOM) National Healthcare Quality Report Categories

IOM Care Need

Living with Illness

IOM Domain

Effectiveness

Identifying Information and Availability

Bibliographic Source(s)

Klings ES, Machado RF, Barst RJ, Morris CR, Mubarak KK, Gordeuk VR, Kato GJ, Ataga KI, Gibbs JS, Castro O, Rosenzweig EB, Sood N, Hsu L, Wilson KC, Telen MJ, DeCastro LM, Krishnamurti L, Steinberg MH, Badesch DB, Gladwin MT, American Thoracic Society Ad Hoc Committee on Pulmonary Hypertension of Sickle Cell Disease. An official American Thoracic Society clinical practice guideline: diagnosis, risk stratification, and management of pulmonary hypertension of sickle cell disease. Am J Respir Crit Care Med. 2014 Mar 15;189(6):727-40. [75 references] PubMed

Adaptation

Not applicable: The guideline was not adapted from another source.

Date Released

2014 Mar 15

Guideline Developer(s)

American Thoracic Society - Medical Specialty Society

Source(s) of Funding

This document was approved and funded by the American Thoracic Society (ATS).

Guideline Committee

American Thoracic Society Ad Hoc Sub-Committee on Pulmonary Hypertension of Sickle Cell Disease

Composition of Group That Authored the Guideline

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*E.S.K. and R.F.M. are co–first authors of this document.
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Financial Disclosures/Conflicts of Interest

All committee members disclosed their potential conflicts of interest to the American Thoracic Society (ATS) and the committee chair. The committee chair reviewed all potential conflicts of interest, discussed them with the chair of the Ethics and Conflict of Interest Committee of the ATS, and resolved them with individual committee members. Committee members were required to refrain from discussing topics related to their potential conflicts of interest both at the face-to-face meetings and in preliminary drafts of the document.

Author Disclosures

R.J.B was a consultant to Actelion ($50,000–99,999), Gilead ($1,000– 9,999), Ikaria $10,000–49,999), Novartis ($10,000–49,999), and Pfizer ($100,000– 249,999); she was a speaker for Actelion ($1,000–9,999). K.K.M. was a consultant to Actelion ($1,000–9,999) and Gilead ($1,000– 9,999) and served on advisory committees of Actelion ($1,000–9,999) and Gilead ($1,000– 9,999); he was a speaker for Actelion ($1,000–9,999) and Gilead ($1,000–9,999), and received research support from Actelion ($25,000–49,999), Fibrogen ($25,000–49,999), Gilead ($25,000–49,999), Novartis ($25,000– 49,999), and Pfizer ($25,000–49,999). G.J.K. received research support from Ikaria ($100,000–249,999). K.I.A. received research support from BioMarin ($25,000–49,999), HemaQuest ($100,000–249,999), Eli Lilly ($25,000–49,999), and TRF Pharma ($50,000–99,999). J.S.G. served on advisory committees of Actelion ($1,000–4,999), Bayer Schering ($1,000–4,999), GlaxoSmithKline ($1,000–4,999), Pfizer ($1,000–4,999), and United Therapeutics ($1,000–4,999); he was a speaker for Actelion ($1,000–4,999), GlaxoSmithKline ($1,000–4,999), Lilly ($1,000– 4,999), Pfizer ($1,000–4,999), and Schering ($1,000–4,999); he received research support from BioMarin ($10,000–49,999) and Lung Rx ($1,000–9,999). O.C. was a consultant to Actelion ($1–4,999), Icagen ($1–4,999), and Cellerant ($1–4,999). E.B.R. was on advisory committees of Actelion ($5,000–24,999) and United Therapeutics ($5,000–24,999), and a speaker for Actelion ($5,000–24,999), Gilead ($5,000–24,999), and United Therapeutics ($5,000–24,999); she received research support from Actelion ($50,000–99,999), Gilead ($50,000–99,999), Lilly ICOS ($25,000– 49,999), and United Therapeutics ($50,000– 99,999). L.H. was a consultant to BioMarin ($1–4,999) and Eli Lilly ($1–4,999), and received research support from BioMarin ($50,000–99,999), Ikaria/INO Therapeutics ($50,000–99,999), and GlycoMimetics ($25,000–49,999). K.C.W.'s spouse previously held stocks or options of Moody Lynn and Co. and State Street Bank ($50,000–99,999). M.J.T. was a consultant to GlycoMimetics ($25,000–49,999) and received research support from Dilafor ($25,000–49,999). L.M.D. received research support from Anthera Pharmaceuticals ($1,000–4,999) and GlycoMimetics ($1,000–4,999). L.K. received research support from INO Therapeutics/Ikaria ($50,000–99,999). M.H.S. was a consultant to Ikaria ($1–4,999) and served on an advisory committee of TRF Pharma ($1–4,999). D.B.B. was a consultant to Actelion ($25,000–49,999), Arena ($1–4,999), Bayer ($5,000–24,999), Gilead ($5,000–24,999), Ikaria ($1–4,999), Lung Rx ($1–4,999), Pfizer ($5,000– 24,999), and United therapeutics ($1– 4,999); he was on an advisory committee of MondoGen ($1–4,999), and received research support from Actelion ($25,000–49,999), Bayer ($100,000–249,999), Gilead ($100,000–249,999), Ikaria ($1–4,999), Novartis ($50,000–99,999), and United Therapeutics ($100,000–249,999). M.T.G. (Co-Chair) received research support from Bayer (USA) ($10,000–49,999) and Gilead ($100,000–249,999); he received royalties from a patent for the use of nitrite salts for cardiovascular indications ($1,000– 4,999). E.S.K., R.F.M., C.R.M., V.R.G., and N.S. reported no relevant commercial interests.

Guideline Endorser(s)

American College of Chest Physicians - Medical Specialty Society

Pulmonary Hypertension Association - Disease Specific Society

Guideline Status

This is the current release of the guideline.

This guideline meets NGC's 2013 (revised) inclusion criteria.

Guideline Availability

Electronic copies: Available from the American Thoracic Society Web site.

Availability of Companion Documents

The following is available:

- Klings ES, Machado RF, Barst RJ, Morris CR, Mubarak KK, Gordeuk VR, Kato GJ, Ataga KI, Gibbs JS, Castro O, Rosenzweig EB, Sood N, Hsu L, Wilson KC, Telen MJ, DeCastro LM, Krishnamurti L, Steinberg MH, Badesch DB, Gladwin MT. Diagnosis, risk stratification, and management of pulmonary hypertension of sickle cell disease. Online supplement. 2014 Mar. 49 p. Electronic copies: Available from the American Thoracic Society Web site .

Patient Resources

None available

NGC Status

This NGC summary was completed by ECRI Institute on April 17, 2015. The information was verified by the guideline developer on June 10, 2015.

Copyright Statement

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